A STUDY TO DETERMINE THE ACCURACY OF THE PLACEMENT OF PULL-THROUGH BOWEL IN CHILDREN WITH ANORECTAL MALFORMATION WHO HAVE UNDERGONE SACROPERINEAL PULL-THROUGH AFTER ACCURATE INTRAOPERATIVE LOCALISATION OF SPHINCTER COMPLEX USING MUSCLE STIMULATOR

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CERTIFICATE

This to certify that the dissertation entitled "A STUDY TO DETERMINE THE ACCURACY OF PLACEMENT OF PULL-THROUGH BOWEL IN CHILDREN WITH ANORECTAL MALFORMATION WHO HAVE UNDERGONE SACROPERINEAL PULL-THROUGH AFTER ACCURATE INTRAOPERATIVE LOCALISATION OF SPHINCTER COMPLEX USING MUSCLE STIMULATOR" is the bonafide work of Dr. Saji Vargheese submitted in partial fulfilment of the rules and regulations for the M.Ch. Branch-V (Paediatric Surgery) examination of the Tamil Nadu Dr. M.G.R Medical University, Chennai to be held in August 2009.

Signature :	**Signature :**
Guide:	**Head of the Department**
Dr. Jacob Chacko; M.S., M. Ch., Dip. NB	**Dr. Sudipta Sen M.S, M.Ch.**
Professor	**Professor & Head**

Department of Paediatric Surgery,
Christian Medical College,
Vellore – 632004,
Tamil Nadu, India.

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Contents

1. Introduction

Anorectal malformations (ARM) are relatively frequently encountered anomalies that represent an important component of paediatric surgical practice. Many in our profession have significant interest in the management of numerous variants of ARM.

Malformations range from minor, easily treated defects that carry an excellent functional prognosis, to complex defects that are difficult to treat, are often associated with other anomalies and carry a poor functional prognosis.

Over the last half century or more, the treatment of ARM evolved from a simple cut-back procedure / translocation anoplasty to the abdomino-perineal and later the sacro-perineal pull through procedures. The sacro-perineal pull-through was widely practiced till the introduction of the posterior sagittal anorectoplasty (PSARP operation).

The current goals in the management of these defects are 1) to anatomically reconstruct all malformations, 2) to recognise and treat any associated defects that may be life-threatening and 3) to treat the functional sequeale of the malformations, in order to provide these patients with a good quality of life.

The long term functional outcome after repair of intermediate and high ARM remain far from perfect. Majority of children have significant problems like soiling and constipation in the long term. Despite recent developments in the understanding of the embryology and surgical anatomy of ARM, it remains unclear which is the optimal repair of these complex anomalies.

PSARP although it gives a superior exposure, it involves dividing the sphincter muscles in the midline through it and suturing the muscles back. Whenever the muscles are divided and sutured, there will be healing by fibrosis which will affect the ultimate function of the sphincter muscles

which has been documented by MRI studies. The Stephen's procedure involves blind hooking of the sphincter muscles and therefore it is possible that one may miss part of the sphincter muscles when bringing the bowel down.

In our technique we have been accurately identifying the sphincter muscle complex by intra-operative muscle stimulation and pulling the bowel through it . It is hoped that this will result in a more accurate placement of the bowel within the sphincter and therefore leads to a better functional outcome.

2. Aims

1. To identify the ano-rectal sphincter complex intra-operatively by direct stimulation with muscle stimulator and pulling the rectum through this muscle complex during sacro-perineal pull-through procedures for intermediate ano-rectal malformations.

2. To study the relationships of this pulled-through rectum to the ano-rectal sphincter complex post-operatively by MRI .

3. To assess the efficacy of the ano-rectal sphincter by determining resting pressure and squeeze pressure post operatively by ano-rectal manometry.

4. To determine functional outcome in these patients after colostomy closure

3. Review of Literature

Paul of Aegineta in the seventh century pierced an anal membrane and used a wedge shaped tent dilator (1). In 1576, Galen described the anal sphincters, levator muscles and coccyx (2). Cook, 1676 treated a child by making a small incision over a blind anal membrane and dilated the aperture with an elder pith. He emphasised care of the sphincter muscles to others who sought to duplicate his success (3,4). Saviard was the first to attempt treatment of a high termination of the bowel by plunging a trocar through the perineum (5). In 1787, 94 years later, Benjamin Bell performed the first perineal dissection in 2 new-borns, finding the blind ending rectum at variable lengths from just above the anal area to the level of coccyx (6). A trocar was inserted and fecal content evacuated. In 1792, Mantell reported a girl with recto-vaginal fistula and performed an incision in the perineum and carried it up to a probe placed through the vagina into the fistula creating an anal communication. Re-operation was required 2 years later for anal stricture (7).

The first successful sigmoid colostomy (termed an “inguinal colostomy” or “procedure of Littre”) was performed by Duret in 1793 on a female infant who survived into adult life (8). In 1978, Martin of Lyon suggested insertion of sound in the colostomy and pushing distally to identify the blind ending rectum during a later perineal dissection (9). In 1834, Roux of Brignoles attempted to preserve external sphincter function and used a midline longitudinal incision extended towards the coccyx (10). The incision continued through the elliptical sphincter ani muscle and levators and when the rectal-artesia was palpated, a trocar was inserted into the bowel, releasing the meconium. In 1835, Amussat performed the first proctoplasty by suturing the opened rectal-atresia to the skin in the midline (11). This was a landmark procedure at that time and gained wide acceptance and was used frequently for the rest of the 19th century. Techniques to repair recto-vaginal and recto-vulvar fistulae were described by Dieffenbanch in 1845 (12). In 1860, Bodenhamer detected the presence of sphincter muscles in some instances of high rectal atresia (13,14). Despite this

observation he shunned colostomy and recommended that an artificial anus be always established in the perineum. He championed the midsagittal incision first described by Roux 27 years earlier. In 1879, Mcleod described abdomino-perineal procedure (AP) for instances in which the blind rectal atretic end was not found below (15). In 1886, McCormae was one of the few to suggest a 2-staged procedure – preliminary colostomy and subsequent proctoplasty (16). In 1897, Matas combined a sacral approach to rectal atresia with sacrotomy to avoid exposure in instances of high lying anomalies and predicted that this would the route of choice for these procedures in the future (17).

In 1930 Wangensteen and Rice described the radiographic invertogram as a method of determining the level of termination of rectal atresia and deciding whether a perineal approach was rational (18). Drs. Ladd and Gross at the Boston children's Hospital kept their dissection close to the hollow of the sacrum and the external sphincter was divided in lateral halves and re-sutured in front and behind the proctoplasty (19). Till this time, an early perineal approach was considered the method of choice (20). Following world was II, things began to change. The availability of antibiotics and improvements in anesthesia had a positive influence in reducing the septic complications associated with bowel surgery. In 1948, Rhoads and colleagues in Philadelphia rekindled interest in a combined AP approach for cases of imperforate anus and high rectal atresia (21). Browne popularised the "cut-back" anoplasty for instances of perineal fistula (22). In 1953, Douglas Stephens with Dennis Browne, in London described the sacroperineal rectoplasty and emphasised the role of the levator ani and downplayed the importance of internal and external sphincters (23,24). In 1959, Fritz Rehebein of Bremen re-introduced the endo-rectal pull-through with an abdomino-perineal approach for boys with recto-urethral fistula (25). Rehebein divided the bowel at laprotomy, stripped the mucosa from the distal rectal atretic end and brought the proximal bowel through the resultant muscular sleeve to the anal dimple to peroform an anoplasty (25). He missed the puborectalis sling in performing this procedure. In 1961 after extensive dissections, Stephens

proposed the importance of puborectalis as the main muscle of continence but recognised that soiling accidents continue to occur because of leakage from the anal canal due to total or functional absence of the external sphincter (26). In 1963, because of the high incidence of the incontinence with the AP approach, Kiesewetter of Pittsburg modified Stephen's operation by performing an abdomino sacro-perineal procedure (27). Unlike Stephens, Kiesewetter believed that the external sphincter muscle was present and worth saving . Two years earlier (1961), E. Ide Smith and Robert E. Gross identified a normal-sized external sphincter in 15 patients of 16 autopsies they performed (28). In 1967, Swenson and Donnellan of Chicago described their experience with AP procedures and preservation of puborectalis sling but completely ignored the external sphincter muscle and did not think it was important in controlling continence (29). In 1978, dissatisfied with outcomes of other procedures, Mollard, recommended an anterior perineal approach bringing an atretic bowel down in front of the puborectalis sling (30). However, mucosal prolapse and soiling continued to be a problem. The reuslts of the various procedures were difficult to asses as different subjective criteria for grading and definitions were used by various authors to assess the functions. Incontinece remained a major problem. The posterior sagittal anorectoplasty (PSARP) operation described by Peter de Vries and Alberto Pena and published in 1982 was a new landmark event in the history of ARM (31) and was rapidly adopted by many pediatric surgeons throughout the world. They redefined the arrangement of pelvic muscles and sphincters as a fused sphincter muscle complex. They completely divided all the muscles posteriorly in the midline from the anal dimple to the coccyx. The distal atretic segment was tapered to fit within the puborectalis and the divided muscles were re-sutured posteriorly around and to the neo-rectum prior to performing the anoplasty. Although the rate of continence improved it became apparent that many children had significant motility disorders and fecal retention was a major problem (32,33,34). Heightened awareness of this problem led to the establishment of close follow-up programs to assure patient and parent compliance with post-operative dilatations and appropriate rectal washouts using enema (34,35).

Pena and his associates employed the posterior sagittal approach to repair the cloacal anomalies and develop considerable experience in the management of these patients (36, 37, 38, 39).

In 1992, Malone described the antegrade colonic enema as an alternative to traditional retrograde enema washouts from below in children with incontinence or significant fecal retention (40). In 1990s advances in technology resulted in new methods of assessment of patients with ARM and their associated anomalies. Transperineal ultrasonography was used to locate the infracoccygeal level of atresia and identify fistula (41, 42). MRI imaging proved useful in evaluating the pelvic and perineal muscle status and identifying instances of tethered cord, vertebral anomalies and spinal dysraphic syndromes (43,44,45). Post-operative assessment of sphincter muscle complex and the position of the pulled-thorugh segment within the sphincter complex is assessed by anal-endo-sonography (46,47). Georgeson (48) employed a laparoscopic assisted one-stage AP pull though. Iwanaka et al (49) and Yamataka et al (50) used a laparoscopic muscle stimulator to accurately locate the sphincter during laparoscope assisted repair.

There remains several areas of controversy regarding the choice and timing of the procedure and methodology used to assess results. Designing a protocol that will define the precise location of the pulled-through rectal segment and pelvic and sphincter muscle assessment using MRI and / or anal endo-sonography, anal manometrics, functional defecography and colonic motility studies in addition evaluation of soiling, sensation and other subjective analysis is needed.

4. Anatomy

According to Gray's anatomy (Williams 1996), the large intestine consists of the cecum, colon, rectum and anal canal and its opening on the surface of the perineum is called anus. The anus is located in the middle of the line joining the two ischial tuberosities.

The rectum has been unequivocally described as being continuous above with the sigmoid colon at the level of S3 vertebra , the junction been indicated by the lower end of the sigmoid mesocolon and as being continuous below with the anal canal by passing through the pelvic diaphragm. The ano-rectal junction has been described as being situated 2-3 cm in front of and slightly below the tip of the coccyx, from this level the anal canal passes downwards and backwards from the lower end of the rectum, the backward bend of the gut at the ano-rectal junction being termed the perineal flexure of the rectum.

The rectum is lined by single layer of columnar epithelium with goblet cells, mucous secreting glands and crypts and is capable of absorbing water and crystalloids. The taenia-coli above fuse to give it a complete coat of longitudinal muscle; its coat of circular muscle is also complete and uniform. It is concave, forwards and has three lateral curves. In the adult the upper diameter is 4 cm; it then dilates to an ampulla and then narrows abruptly at its lower end. It consists functionally of two parts, the upper free to distend towards the peritoneal cavity and the lower, enclosed in condensed extra-peritoneal tissue.

The anal canal begins where the lower end of the ampulla of the rectum suddenly narrows and passes downwards and backwards to end at the anus. It is surrounded over its whole length by the sphincter that normally keeps the canal closed. It is 4cm long in adults and its shows in its upper 1.5 cm columns, valves and sinuses. The epithelial lining at its upper end resembles rectum with a single layer of columnar cells and intestinal glands. On the other hand, the lower end is skin-lined

with hair and sweat glands. The junctional area is the pectinate line; above, the epithelium is stratified columnar or transitional or low-stratified polygonal; below it is non-keratinised stratified squamous.

The sphincter muscle complex mechanism is traditionally described as including the voluntary striated muscles of the external sphincter, the levator musculature and the involuntary smooth muscle, the internal sphincter. These muscles are innervated by the pudendal nerve, both motor to the external sphincter and sensory to the skin around the anus and derived from the sacral plexus roots S2, S3, S4 as well as autonomic nervous system via nervi-erigentis also derived from segments S2, S3, S4 segments of the spinal cord.

1. **The external anal sphincter :**

The sphincter has 3 open U-loop like sections which gives it an ellipsoidal shape morphologically (50). It is situated between the fat-filled ischiorectal fossae laterally, it attaches anteriorly to the perineal muscles and posteriorly to the ano-coccygeal raphe and coccyx (51). It is divided into sections that are continuous with one. From caudal to cranial these are:

a. The superficial external sphincter (sub-cutaneous) : This surrounds the lower anal canal and is traversed by the coat tails of the longitudinal anal canal muscles. Histologically it can be seen as 2 parallel muscle strips in the axial plane; confirmed operatively as the so-called parasagittal fibres and by MR imaging.

b. The deep external sphincter : surrounds the middle anal canal anterorly. It is continuous through the high anal canal with the puborectalis fibres of the levator ani cranially; with which seen in the sagittal plane, it gives a posteriorly positioned tear drop-like appearance to the anal sphincters perpendicular to the axis of the anal canal.

c. The puborectalis fibres of the levator ani : Contraction of this section of the levator muscle forms the ano-rectal ring. During contraction, it cannot be separated from the

external anal sphincter caudally, but from the rest of the levator formed pelvic diaphragm cranially. Its lower border defines the upper-extent of the high anal canal. The puborectalis is continuous with, but not described as part of the anatomic external anal sphincter.

2. **The internal anal sphincter :**

This is the terminal portion of the inner-circular smooth muscle layer of the rectum. It is described as being composed of 26 rings, flat ring-like slates of smooth muscle bundles stacked like the slates of a Venetian blind, one on the top of the other and arranged to form 3 equally sized columns around the anal canal (52). It surrounds the upper two-thirds of the anal-canal above the anal valve-line. Its lower border is identified by the inter sphicnteric groove on the skin, thus defining the upper limit of the low-canal zone. Injury during surgery results in incontinence, resulting in reversal of the normal resting pressure in the anal canal (53)

3. **The Levator Ani :**

This is the major muscle of the pelvic diaphragm and is attached anteriorly to the pubic bone. The ventro-medial segment is termed the pubovisceralis muscle as it holds the urethra, the vagina and ano-rectum within its sling-like fibres. It is drawn caudally by the viscera passing through it to which it is attached. A further sub-division, a segment composed of fibres passing, but intimately in contact with the ano-rectum in the shape of a U-loop from pubis to pubis is named purborectalis. It forms the ano-rectal junction defining the rectum from the surgical anal canal the ano-rectal ring. This muscle participates in the formation of the external anal sphincter.

5. Physiology

Mechanism of Defecation

Various reflexes such as the gastro-colic reflex and the ileo-colic reflex, contraction of the colon, caused by filling the stomach and the ileum respectively as well as voluntary contraction of the abdominal musculature may initiate defecation by suddenly filling the rectum with colonic contents. The increasing intra-rectal pressure stimulates the distension receptors in the puborectalis muscles and the para-puborectal tissues and the desire to pass stool is consciously felt. This allows even the smallest amounts of stool to reach the anal canal. The hypersensitive mucosa of the anal canal in the region of the anal walls is able to discriminate between flatus and liquid or solid stools. The reflex contraction of the external anal sphincter and the pubo-rectalis will prevent the expulsion of stools from the anal canal and thus inhibit fecal soiling. This effect is increased by the compression of the lower anal canal by the corpus cavernosum of the rectum and by the corrugator muscle of the anus. This allows the rectum time to adapt itself to the increased intra-luminal pressure. The aboral-oral pressure gradient of the rectum will propel the stools upwards into a more proximal rectal segment. This, however will stimulate further propulsive waves via a feedback mechanism. An intra-rectal pressure between 25-30mm Hg will stimulate a reflex inhibition of the anorectal sphincters and the puborectalis muscle. The voluntary contractions of the abdominal muscle will also cause reciprocal inhibition of the striated muscle of the pelvic floor. This, in turn will decrease the acuteness of the ano-rectal angle formed by the puborectalis muscle and defecation commences. When the recto-anal reflex operates, following sudden distension of the rectum, sampling of whether the waste is solid, liquid or gas occurs at approximately at the level of the anal valves. If the defecation is not intended, voluntary contraction of the puborectalis muscle will return the contents back into the rectum off the sensitive zone and the desire to defecate will diminish.

The external sphincter is a powerful muscle brought into action in moments of stress to supplement the sling action in arresting defecation or deflation. It too has a resting tone that mildly occludes the anus, and when forced open by flatus under high pressure it exhibits a flutter valve action with the accompanying characteristic noise. The tone of the internal and external sphincters that surround the skin line anal canal is probably responsible for prevention of wetting of this part of the anal canal with mucous secreted from the adjoining rectal mucosa in the long intervals between the acts of defecation. Neither of these sphincters accounts for minute-to-minute day and night fecal continence, which appears to be the function of the sleeve and sling.

Children who became chronically constipated indicate that the sleeve and sling become easily tired by the impacting faeces, become relaxed and permit shortening of the anal canal to the length only of skin lined anus. It is found that then the short passage although encircled by external and internal sphincters is barely sphincteric, permitting constant leakage, which is momentarily arrested only at the time of conscious muscular contractions of the external sphincter surrounding the skin-lined anus.

Finally, the anal canal constructed in patients exhibiting a congenital recto-urethral fistula is endowed with a high degree of sensation, content discrimination and muscular sphincter function if the new canal is lodged within the striated muscle complex which then is its only sphincter. If the canal is directed to the perineum through the muscular diaphragm posterior to the sling, the bowel lacks appreciation of its content and all power to control defecation (27, 54, 55).

Duthrie and Gairus (56) believed that a skin-lined canal is vital to continence. Kieswetter and Nixon (57) showed an ingrowth of sensory fibres from the perineal skin following pull-through operation. Anoplasty to create a skin-lined canal if prolapse occurs after pull-through operations improves continence (58). Stephens & Smith (59), considered that proper function of the puborectalis muscle is adequate for near complete continence including content discrimination, as is found in patients after rectoplasty operations for ARM.

6. Classification

ARM represents a wide spectrum of defects and conditions. A clear understanding of the normal ano-rectal anatomy and the different types of ARM is necessary for both the planning of the surgery and the procedure itself. An appreciation of the classification systems is useful in practice to the surgeon.

Amussat, in 1835 was the first to attempt a classification system of ARM and described 5 groups (60). In 1934 Ladd and Gross proposed a classification system which became the standard (Table 1).

Table 1 – Ladd and Gross Classification (61)

Type	Anomaly
I	Anal & anorectal stenosis
II	Imperforate Anus
III	Imperforate Anus with blind ending pouch with fistula
IV	Rectal atresia

1n 1963, a Melbourne team led by Stephens classified the lesions into 2 categories either high or low (table 2.). This classification recognises the importance of puborectalis muscle and its effects in continence. Lesions above pubococcygeal line (PC) were described as high and below as low. The PC line is drawn on a lateral pelvic radiograph “invertogram” between the midpoint of the pubis and the inferior aspect of the sacrum (18). It represents the level of Levator Ani attachment to the pelvic wall.

Table 2 – Stephens and Smith 1963 Classification Based on Embryological Concepts

Anorectal Deformities	
A. Defect of the partition of the internal cloaca	
Male	Female
1. Anorectal agenesis (no fistula)	1. Anorectal Agenesis (no fistula)
2. Rectovesical fistula	2. Rectovesical fistula
3. Rectourethral fistula	3. Rectourethral fistula
	4. Rectovaginal fistula
	5. Rectovestibular fistula
B. Secondary Defects After Partition	
1. Rectal Atresia	1. Rectal Atresia

Rectal Deformities	
A. Defect of the perineum	
Male	Female
1. Anterior perineal anus	1. Anterior perineal anus Perineal Vulvar
	2. Anovestibular fistula
	3.Perineal groove
	4. Perineal canal
B. Defects of the Genital Fold	
1. Covered anus complete	1. Covered anus complete
2. Ano cutaneous fistula	2. Ano cutaneous fistula
3. Ano bulbar fistula	3. Ano vulvar fistula
C. Defects of the Proctodeal Pit	
1. Anorectal agenesis	1. Anorectal agenesis
2. Imperforate anal membrane	2. Imperforate anal membrane
2. Anal stenosis i. covered anal stenosis	3.Anal stenosis i. covered anal stenosis

ii. anal membrane stenosis iii. anorectal stenosis	ii. anal membrane stenosis iii. anorectal stenosis
D. Unclassified	
1. Vesico-intestinal fissure	
2. Duplication of the rectum and anus	
3. Combination of usual deformities	

In 1964, Santulli proposed his classification system which was based on the work of Ladd and Gross. This also divided lesions into low infra-levator and high supra-levator (Table 3) (62).

Table 3: Anorectal anomalies based on a simplified Santulli Classification

In 1970, the "International" classification was proposed at a symposium on ARM at the pediatric surgical congress in Melbourne in order to further decrease confusion (table 4). Based on the earlier work of Smith and Stephens, the 1970 international classification was based on the principles of normal and abnormal anatomy and divided the lesion into 3 groups – high supra-levator, intermediate and low (trans levator). Although it proved much too complex for most surgical groups due to the facts, it contained nearly 40 sub-types. It is still in use in the literature.

Type of Anomaly	Female	Male
Low Infra-levator	1. Anal stenosis 2. Anal membrane 3. Anal agenesis a. With fistula b. Without fistula	1. Anal stenosis 2. Anal membrane 3. Anal agenesis a. With fistula b. Without fistula
High, supra levator	1. Rectal agenesis a. With fistula b. Without fistula 2. Rectal atresia	1. Rectal agenesis a. With fistula b. Without fistula 2. Rectal atresia

Type of Anomaly	Female	Male
High	1. Anorectal agenesis a. Rectal atresia b. With fistula Rectocloacal fistula Rectovaginal / high 2. Rectal atresia	1. Anorectal agenesis a. Rectal atresia Rectovesical fistula Rectourethral fistula 2. Rectal atresia
Intermediate	1. Anal agenesis a. Without fistula b. With fistula Rectovaginal fistula low Rectovestibular fistula 2. Anorectal stenosis	1. Anal agenesis a. Without fistula b. With fistula Recto bulbar fistula 2. Anorectal stenosis
Low	1. At normal anal site Covered anus complete Covered anal stenosis 2. At perineal site Ano cutaneous fistula Anterior perineal anus 3. At vulvar site Vulvar anus Ano vulvar fistula Ano vestibular fistula	1. At normal anal site Covered anus complete Covered anal stenosis 2. At perineal site Ano cutaneous fistula Anterior perineal anus
Miscellaneous	Anal membrane stenosis Imperforated anal Membrane Perineal groove Perineal canal	Anal membrane stenosis Imperforated anal membrane Perineal groove Perineal canal

Table 4: A simplified version of the International Classification.

New research and variations in surgical technique in the late 1970s and early 1980s altered previously fixed concepts. This led to the “Wingspread” classification, which evolved from a conference held in the Wingspread Convention Centre Racine, Wisconsin, USA in 1984 (63). It was created in order to update the “international” Melbourne classification that was described at that time as “unwieldy”. It is based on high, intermediate & low anomalies and is presented in Table 5.

Table 5. Wingspread Conference Classification

Level of Anomaly	Male	Female
High	1. Anorectal agenesis a. Rectovesical fistula b. Without fistula 2. Rectal atresia	1. Anorectal agenesis a.Rectovaginal fistula b.Without fistula 2. Rectal atresia
Intermediate	1. Rectourethral fistula 2. Anal agenesis without fistula	1. Rectovestibular fistula 2. Rectovaginal fistula 3. Anal agenesis without fistula
Low	1.Anocutaneous (perineal fistula) 2. Anal stenosis	1. Ano vestibular fistula (perineal fistula) 2. Anocutaneous (perineal fistula) 3. Anal stenosis
Miscellaneous	Rare malformation	Persistent cloacal anomaly Rare malformation

However the Wingspread classification was not fully endorsed by the surgical community as it is based on anatomical principles. Alberto Pena proposed a classification based on the anatomical defect and how they correlate with surgical management (Table 6)

Table 6: Pena'sClassification

Male	Female
Perineal (cutaneous) fistula	Perineal (cutaneous) fistula
Rectourethral fistula Bulbar Prostatic	Vestibular fistula
Rectovesical fistula	Persistent cloaca
Imperforate anus without fistula	Imperforate anus without fistula
Rectal atresia	Rectal atresia

In May 2005, an international congress for the development of standards for the classification, treatment & follow up of ARM took place in Krickenbeck ,Castle in Westphalia, Germany. At this meeting a new unifying international classification system was introduced which will enable standardisation of definition and treatment protocols for various anomalies. This was known as the Krickenbeck classification (64) .It is not based on the anatomical , embryological or on imaging. It is divided into two main groups " major clinical groups" and " rare/ regional variants " and is based on frequency of occurrence and allows management outcomes to be measured (Table 7) .

Table 7 : Standards for diagnostic procedures : International classification (Krickenbeck)

Major clinical groups	1.Perineal (cutaneous) fistula 2.Rectourethral fistula *Bulbar *Prostatic
	3.Rectovesical fistula 4.Vestibular fistula 5.Cloaca 6.No fistula 7.Anal stenosis
Rare/ regional variants	1.Pouch colon 2.Rectal atresia/ stenosis 3.Rectovaginal fistula 4.H type fistula 5.others

Besides a new international “Krickenbeck” standards for diagnostic procedures, an international grouping of surgical procedures for follow up was developed at the Krickenbeck meeting . This second standardisation seemed to be necessary to make the different surgical procedures comparable with each other (65) (Table 8).

Table 8: International grouping (Krickenbeck) of surgical procedures for follow-up.

Operative procedures:	Perineal operation. Anterior sagittal approach. Sacroperineal procedure. PSARP. Abdomino-sacroperineal pull-through. Abdominoperineal pull-through. Laparoscopic assisted pull-through.
Associated conditions :	Sacral anomalies. Tethered cord.

7. Scoring Post-operative Results

Standardized assessment of clinical outcome after repair of ARM is essential for appropriate quality control and for comparing different treatment modalities. Clinical assessment is subjective and may be biased by the observer who is often the surgeon treating the patient .Therefore scales and scores that provide reliable information on the condition and functional status is needed . However , appropriate methods and instruments for collecting data on the outcome after repair of ARM have been a matter of debate for decades .

A scale is an instrument that is used to measure clinical phenomena , such as degree of incontinence or the squeezing pressure of the anal sphincter . A score is a value on a scale in a given patient. Scores in specific patient may be dichotomous (yes/no) or value ordered .Thus qualitative scores can be differentiated from numerical scores. Principally a score may serve three functions: prediction, evaluation over time or description at a certain time point(66) , Scores have to be reproductive, valid and responsive. Ideally the process of ensuring reproductibility, validity or responsiveness of a specific score or score should not be based on the observers clinical knowledge and common sense , but on a structured scored descriptively . None of the scores suggested for validation process concerning reproducibility, validity or responsiveness. In addition, the problem of definition of end points like constipation, intermittent soiling or other symptoms are not uniformly defined .

Specific scores used in patients with ARM :

There is consensus that fecal continence represents the most important endpoint in patients with ARM . Therefore, specific scores for assessment of long term results are focussed on differentiating various degrees of fecal incontinence. No consensus has been achieved on including and scoring other symptoms such as constipation , urinary incontinence , electromanometeric and endosonographic findings or quality of life measurement .

1. The Scott score (67) :

In 1960, Scott established a qualitative score that differentiated between “good” ”fair” , “poor” continence . The items used are defecation habits, stool control, perineal soreness and the function of puborectalis muscle on digital examination. The score was not validated and a clear definition of specific items was not given .

2 .The Kelly score (68) :

The criteria are some what similar to Scott score, but continence is scored qualitatively . The determination is based on leakage phenomenon, on the strength of the puborectalis sphincter and sensitivity . Factors include the appearance of staining or smearing , accidental defecation or soiling , sensitivity, the strength of the puborectalis muscle action on digital examination and “ feeling of defecation”. A total of 5-6 points is considered – good , 3-4 points – fair and 2 points – poor . It is probably the most commonly used instrument for assessment of fecal incontinence today . It is not often used as a single instrument , but is compared with other more objective measures such as manometery, electromyography and quality of life data (Table 9).

Table 9: Kelly score

Staining / Smearing	More 2 Occasional 1 Constant 0
Accidental defecation / soiling	More 2 Occasional 1 Constant 0
Strength of the puborectalis muscle	Strong 2 Weak 1 None 0

3. The Holschneider score (69) :

Holsehneider and Metzer introduced a qualitative clinical score , including the parameters, frequency of defecation, fecal consistency , soiling ,rectal sensation, ability to hold back, discrimination & need of therapy. Each of these parameters is scored as 0-2 according to degree of impairment. A score of 10-14 is “continent” ,”5-9” points “fair’ & 0-4 points “incontinent”. Later the score was modified by reducing clinical parameters & including manometric data without changing the numerical scoring (Table 10)

Table 10: Holschneider score (1983)

Frequency of defecation	Normal (1-2/day)	2
	Often (3-5/day)	1
	Very often	0
Fecal consistency	Normal	2
	Soft	1
	Liquid	0
Soiling	No	2
	Stress/ diarrhoea	1
	Constant	0
Sensitivity	Normal	2
	Reduced (no discrimination)	1
	Missing	0
Anorectal resting pressure profile	≥ 20-24 mmHg	2
	14-19mmHg	1
	˂13mmHg	0
Maximum pressure at maximum squeezing	≥30mmHg	2
	20-29mmHg	1
	˂20mmHg	0
Adaptation reaction	Normal	2
	Small amplitude shortened	1
	Not detectable	0

4 . The Wingspread Score (70) :

The grades of continence are scored qualitatively . They fall into 4 main categories of “clean” , staining” , “ intermittent soiling”, & “constant fecal soiling “. Subcategories include the need of occasional or constant therapy. In an additional category , related complications concerning the anorectum, urinary, genital, & other functions are noted.

Table 11: Wingspread score according to Stephen et al (1988)

1. Clean

1.1 No accumulated feces

1.11 No therapy.

1.12 Occasional therapy.

1.13 Therapy dependent.

1.2 Accumulated feces

1.21 No therapy.

1.22 Occasional therapy.

1.23 Therapy dependent.

2 . Staining

2.1 No accumulated feces

2.11 No therapy

2.12 Occasional therapy.

2.13 Therapy dependent

3 . Intermittent fecal soiling

3.1 No accumulated feces

3.11 No therapy

3.12 Occasional therapy

3.13 Therapy dependent

3.2 Accumulated feces

3.21 No therapy

3.22 Occasional therapy

3.23 Therapy dependent

4 .Constant fecal soling

4.1 No accumulated feces

4.11 No therapy

4.12 Occasional therapy

4.13 Therapy dependant

4.2 Accumulated feces

4.21 No therapy

4.22 Occasional therapy

4.23 Therapy dependant

Related complications (specify)

1. Anorectal

(a) Abnormal position

(b) Stenosis

(c) Prolapse

(d) fistula.

(e) lack of contractility.

(f) Abnormal length

2. Urinary

3. Genital

4. Other

5 .The Rintala Score :

Rintala and Lindahl (70) established a clinical score for the evaluation of fecal continence. The score is derived from standardized questionnaires and physical examination is not required. The score consists of seven factors, which are scored from 0 to 3, except the factor of frequency of defecation which is scored 1-2. The authors initially compared the scores of ARM children with normal controls, The score of 18 or above is taken as normal with “excellent” outcome after repair. The group with scores 9-16 was with “good” results having occasional staining and infrequent accidents. Patients with “fair” results had intermittent daily soiling or staining and scored 7-11 points. Patients with “poor” results scored 6-9 points and had to use daily enemas because of severe constipation or had constant soiling. There were same validation steps. The scores derived from the questionnaires and clinical outcome noted in the hospital records were positively correlated. A pathological finding in plain spinal radiograph or MRI negatively correlated with bowel function score. Manometry did not differentiate with excellent and good clinical outcome, but showed a significantly reduced anal resting pressure in patients with fair or poor clinical outcome.(Table 12).

Table :12 Clinical Score for evaluation of fecal continence by Rintala &Lindahl

Ability to hold back defecation	
Always	3
Problems lesser than 1 per week	2
Weekly problems	1
No voluntary control	0
Feels /reports the urge to defecate	
Always	3
Most of the time	2
Uncertain	1
Absent	0
Frequency of defecation	
Every other day to twice daily	2
More often	1
Less often	1
Soiling	
Never	3
Staining less than 1 per week, no change of underwear required	2
Frequent staining and change of underwear required	1
Daily staining requires protective aid	0
Accidents	
Never	3
Fewer than 1 week	2
Weekly accidents, often requires protective aids	1
Daily, requires protective aid during day and night	0
Constipation	
No constipation	3
Manageable with diet	2
Manageable with laxative	1
Manageable with enemas	0
Social problems	
No social problems	3
Sometimes (foul odour)	2
Problems causing restricting in social life	1
Severe social or psychological problems	0

6. Pena 1995 : Pena (71) suggested a specific methodology for evaluation of long-term results according to his personal experience. At the time of evaluation, none of the patients were allowed to be subjected to any type of medical management. Four parameters are evaluated (Table13) :

1) voluntary bowel movement, which are defined as feeling to urge to use the toilet to have a bowel movement, the capacity to verbalize it and to hold the bowel movement.

2) soiling is defined as involuntary leaking of small amount of stool, which may be present with or without voluntary bowel movement. Soiling Grade 1 occurs occasionally (once or twice per week). Grade 2 refers to soiling that occurs everyday, but does not cause social problems. Grade 3 represents constant soiling with social problems.

3) constipation: defined as the incapacity to empty the rectum spontaneously without help everyday. Grade 1 when the patient is manageable by diet, Grade 2 when he requires laxatives, Grade 3 when he requires enemas.

4) urinary incontinence is considered Grade 1 when the patients has mild dribbling and wetness of the underwear day and night, Grade 2 when he is completely incontinent. Patients with voluntary bowel movement and no soiling are considered totally continent.

Table 13:Evaluation of bowel function according to Pena

1. Voluntary bowel movements

Feeling of urge

Capacity to verbalize

Hold the bowel movement

2. Soiling

Grade 1 Occasionally (once or twice per week)

Grade 2 Every day, no social problem

Grade 3 Constant, social problem

3. Constipation

Grade 1 Manageable by changes in diet

Grade 2 Requires laxatives

Grade 3 Requires enemas

4. Urinary incontinence

Grade 1 Mild dribbling / wetness day and night

Grade 2 Complete incontinence

7. Krikecnbeck Scoring (2005):

At the Krickenbeck Meeting in 2005, consensus was achieved concerning the assessment of outcome after anorectal malformation repair. The method comprise three parameters: voluntary bowel movements (yes/no) , soiling (yes/no), soiling ,if yes grade 1-3) and constipation (yes/no, if yes grade 1-3). The assessment should be performed in children more than 3 years of age who are not undergoing therapy. (Table 14).

Table 14: Krickenbeck Scoring 2005

1. Voluntary bowel movements **yes/no**

Feeling of urge

Capacity to verbalize

Hold the bowel movements

2. Soiling **yes/no**

Grade 1 Occasionally (once or twice per week)

Grade 2 Every day, no social problem

Grade 3 Constant, social problem

3. Constipation **yes/no**

Grade 1 Manageable by changes in diet

Grade 2 Requires laxatives

Grade 3 Requires enemas

8. Other scoring systems :

Few objective methods of scoring are used. Holschneider (72) electromanometrically defined grades of continence. He included selected manometric data in his clinical score for obtaining more objective results. Diseth and Emblem (73) confirmed that anal canal resting pressure and squeeze pressure correlated negatively with fecal incontinence. Rintala et al (74) found that manometric parameters that correlated with continence outcome was voluntary squeeze pressure. Fukata et al (75) compared endosonography and electromyography of the external anal sphincter with electromanometry and clinical data derived from the Kelly score. Endosonographic findings for the external anal sphincter corresponded well with electromyographic findings and not with manometry. Jones at al (76) compared endosonography with magnetic resonance imaging after repair of ARM. The findings were comparable in only 9 out of 14 patients. Fukaya et al (77) compared MRI with clinical assessment on the basis of Kelly score. The proportion of “fair” or “poor” developed muscles was not significantly different between the continence groups according to Kelly.

8. Results following treatment of ARM

1 . Operative complications :

A neonatally performed colostomy carries a high degree of morbidity. The most common complications are colostomy and prolapse (78, 79). Stoma complications appear to be less common with a completely divided sigmoid colostomy than with transverse or loop colostomy (80, 81). The reported total incidence of complications of infant colostomies range between 17 and 68% of cases.

Early complications occur following all commonly used reconstructions. Peritonitis, retraction or dehiscence of the pull-through segment and refistula between the bowel and urogental tract are typically severe early complications. The incidents of these major complications range between 10-30% following abdominoperineal or sacroabdominoperineal pull-through (82, 83, 78). Severe complications seem to be less common following posterior sagittal anorectoplasty. In the large series of Pena, serious complications requiring major reoperative surgery occurred in 2% of the cases, mainly following repair of the cloaca (71).

Post-operative anal complications have been common following pull-through operations. Anal stenosis and mucosal prolapse have been found in 15-78% of patients (82, 83, 84, 85). Stenosis has usually been attributed to inadequate anal dilatation during the follow-up period. Anal stenosis may respond to dilatation ; in refractory cases, surgical excision of scar tissue needed. Mucosal prolapse usually requires operative treatment to reduce mucosal soiling and to improve sensation in the neo-anal canal. On the other hand, local anal problems have been rare following posterior sagittal anorectoplasty. Pena reported very few local complications in his series of 798 patients (71).

2 . Urological complications :

Lesions in the urinary tract may complicate surgery for ARM. Infection may occur after definitive repair and is caused in most cases by a urological anomaly, vesicoureteral reflux being the most common (86). Urinary infections may be caused by a rectourinary fistula remnant that is too long (87). Damage to the pelvic innervation and ureter during the dissection of rectal blind pouch may cause urinary incontinence or urethral stricture (87). The incidence of urological injuries associated with surgery are strongly related to the experience of the surgeon (88). A detailed imaging of the rectourinary communication by contrast studies may decrease the possibility of injury. Many of the functional urinary abnormalities previously attributed to surgical intervention are congenital (89,90).

Prognostic factors determining outcome in high and intermediate anomalies

1. The level of rectourogenital connection: is an important prognostic factor of bowel function. Males with a bladder-neck fistula and females with a high confluence cloaca have worse prognosis than patients with a lower connection. The obvious cause of poorer prognosis in high anomalies is the more marked hypoplasia of the voluntary sphincter muscles (71).

2. Sacral abnormalities: severe sacral abnormalities adversely affect long-tern functional outcome. More than two missing sacral vertebral or other major sacral deformities such as hemivertebrae and vertebral fusions, worsen the functional outcome when compared with patients with a more normal sacrum (71, 70). The poor outcome is usually related to sphincter insufficiency. Sacral dysplasia may also cause severe constipation by impairing rectal sensibility (70, 91). Modern imaging like ultrasound and MRI has picked up lot of occult

myelodysplasia in patients with ARM. But the impact of these lesions in long-term bowel function is unclear.

3. Functional role of internal sphincter: the functional role of the internal sphincter following repair of ARM is controversial. The functioning internal sphincter can be demonstrated by the presence of rectoanal relaxation reflex (82, 92, 93). In patients in whom the rectourogenital connection has been preserved at the anorectal repair, a functional internal sphincter has been demonstrated in 40-80% by demonstrating the rectoanal relaxation reflex. The presence of internal sphincter has been shown to correlate with favourable functional outcome (94, 95, 96, 97, 98, 99).

4.Abnormal colonic motility: usually presenting as constipation has been reported to be problem in patients with low ARM and in females with vestibular fistula (82, 71). Following PSARP for higher anomalies, chronic constipation is one of the main functional complications encountered. The incidence of constipation following PSARP procedure varies between 10% (100) and 73% (94, 101). Constipation seems to be more common when internal sphincter preserving techniques have seen used (94, 98, 101). The cause of constipation in unclear; extensive mobilization of the anorectum causing partial sensory denervation of the rectum and impair rectal sensation; rectosigmoid hypomotility (71). Segmental colonic transit time studies in patients with ARM has shown that those with low anomalies have rectosignoid hypomotility, whereas those with high anomalies have generalized colonic motility disturbance (102).

5. Surgical techniques: is a significant prognostic factor. However, this is very difficult to prove since there are no randomized controlled studies. Kiesewetter and Chang (103) found abdominoperineal pull-through to be slightly better than sacroabdominoperineal operations in a

series of 70 patients. . Holschneider at al (104) reported significantly better continence outcome in 21 patients who has PSARP compared with a historical cohort of 16 patients who underwent abdominoperineal pull-through. Templeton and Ditesheim (105) reviewed the outcomes in several series and suggested that the use of full-thickness terminal bowel (abdominoperineal pull-through, sacroperineal pull-through) give better long-term outcome than anorectal pull-through procedures. deVries (106) could not find evidence to support the superiority of any procedure.

6. Timing of surgery: the age of the patients at the time of repair has been suggested to influence the long-term functional outcome. Neonatal abdominoperineal reconstruction was popularized by Rhodes et al in 1948 and was popular in 1950s and 1960s. But many surgeons were not satisfied with the functional results and so started doing staged procedures (107, 108). Recently, early repair during the first 3-6 weeks of life is recommended and some surgeons advocate neonatal surgery. The critical anal dilatations are easier to perform in an infant and it allows the early development for neural pathways between the anal canal and brain facilitating better anorectal sensation and sphincteric function. However at present, there is no clear evidence that neonatal pull-through procedures gives a better functional outcome than surgery done at 6-12 months of age (109).

Long-term bowel function during childhood

Reported long-term functional outcomes in patients with high ARM are highly variable. Most series grade the results as good, fair or poor. The good outcome does not mean that the patient has a normal bowel function. Patients with good results are usually been considered socially continent,

which implied that the defects in bowel function do not cause significant social disability. The Table 15 shows the results by various surgeons before the era of PSARP.

Table 15 : Functional outcome before the era of PSARP

	N	Good	Fair	Poor
Partridge & Gough (110)	63	33%	43%	24%
Trusler & Wilkinson (111)	15	26%	20%	54%
Stephens & Smith (78)	25	56%	32%	12%
Taylor at al (112)	45	24%	20%	56%
Cywes at al (113)	38	42%	35%	23%
Smith et al (114)	18	6%	28%	66%

There are few reports concerning long-term functional outcome following PSARP and the results have been inconsistent (Table 16). Some surgeons report a dismal outcome, with most patients requiring adjunctive measures to maintain social continence. On the other hand, in the series of Pena, approximately one-third of the patients with high or intermediate anomalies could be considered as totally continent.

Table 16: Functional outcome during childhood – high malformations: PSARP

	Total continence	Significant soiling	Constipation
Pena (71)	36%	41%	43%
Rintala & Lindahl (94)	35%	30%	60%
Langemeijer & Molenaar (115)	7%	56%	50%
Rintala & Lindahl (101)	50%	22%	9%

The following results (Table 17) from follow-up studies by Pena and Marc Levitt in children who had repair for ARM in terms of voluntary bowel movement, continence and constipation (116)

Table 17: Results of series by Pena

Types of malformation	VBM	Total continence*	Constipation

Rectal atresia/stenosis	100%	88%	57%
Perineal fistula	100%	100%	50%
Vestibular anus	92%	55%	61%
Imperforate anus with no fistula	89%	52%	55%
Bulbar urethral fistula	81%	31%	59%
Short cloaca	79%	28%	39%
Prostatic fistula	73%	20%	45%
Long cloaca	55%	17%	48%
Bladder neck fistula	35%	0%	15%
Series average	77%	39%	48%
VBM : voluntary bowel movement * Voluntary bowel movements and no soiling			

9. Fecal continence

Most patients who undergo repair of an anorectal malformation suffer from a degree of functional defecating disorder and all suffer from abnormality in their fecal continence mechanism. Approximately 25% of patients are deficient enough in these mechanisms that they are fecally incontinent and cannot have a voluntary bowel movement. Fecal incontinence represents a devastating problem that often prevents a person from becoming socially acceptable which in turn provokes serious psychological sequelae.

Fecal continence depends on three main factors (1) voluntary sphincter muscles (2) anal canal sensation (3) colonic motility.

(1) Voluntary sphincter muscles: are represented by the levators, the muscle complex and external sphincter. Patients with ARM have abnormal voluntary striated muscles with different degrees of hypodevelopment. Voluntary muscle can be used only with the information derived from an intact anal sensory mechanism which many children with ARM lack.

(2) Anal canal sensation: exquisite sensation in normal individuals resides in the anal canal. Except for patients with rectal atresia most patients with ARM are born without an anal canal; therefore, sensation either does not exist or is rudimentary. Patients can perceive distensions of rectum only if rectum has been properly located within the muscle structures. This sensation seems to be a consequence of stretching of the voluntary muscle (proprioception). As liquid stool or soft fecal material does not distend the rectum no sensation may be felt by the patient. Thus to achieve some degree of sensation a bowel control, the patient must have the capacity to form solid stool.

(3) Bowel motility: perhaps the most important factor in fecal continence is bowel motility. In normal individual the rectosigmoid remains quiet for variable periods of time depending on specific defecation habits. The peristaltic contraction of the rectosigmoid that occurs prior to defecation is normally felt by the patient. The normal individual can voluntarily relax the striated muscles, which allows the rectal contents to migrate down into the highly sensitive area of the anal canal. There accurate information is provided concerning the consistency and quality of stool. The voluntary muscles are used to push the rectal contents back up to into the rectosigmoid and to hold them, if desired, until the appropriate time for evacuation. At the time of defecation, the voluntary muscle structures relax.

The main factor that provokes the emptying of the rectosigmoid is a massive involuntary peristaltic contraction. Most patients with ARM suffer from disturbance of this sophisticated bowel motility mechanism. Patients who have undergone PSARP or any other type of sacroperineal approach, in which most distal part of the bowel was preserved (megarectum) show evidence of an over efficient bowel reservoir. The main clinical manifestation of this is constipation, which seems to be more severe in patients with lower defects. When the constipation is not aggressively treated this results in further dilatation of the ectatic rectosigmoid and worsening constipation, The enormously dilated rectosigmoid with normal ganglion cells behaves like a myopathic type of hypomotile colon (34). In these patients who are incontinent, a daily enema successfully cleans the bowel (3%). However, patients who have their reservoir and behave like a perineal colostomy. In these individuals a daily enema with a constipating diet and medications to slow down the colonic motility is indicated.

Bowel management program (BMP)

The bowel management program consists of teaching the patient or his/her parents how to clean the colon once daily so as to stay completely clean in the underwear for 24 hours. The program, although simplistic, is implemented by trial and error over a period of 1 week.

It is important to differentiate real fecal incontinence from overflow pseudoincontinence. In patients with real fecal incontinence, the normal mechanism of bowel control is deficient. In overflow pseudoincontinence patients behaves like they are fecally incontinent but really have severe constipation and overflow. It is extremely important to distinguish between the two in order to identify the origin of the problem and consequently to plan the best treatment.

Of all children with ARM who have undergone correct and successful operation, 75% have voluntary bowel movement after the age of 3 years (71). About half of these patients soil their underwear on occasion and these episodes of soiling are usually related to constipation. Approximately 40% of all these children have voluntary bowel movement and no soiling. Children with good bowel control may still suffer from temporary episodes of fecal incontinence, especially when they experience severe diarrhoea. Some 25% of all children suffer from real focal incontinence and these are the patients who must receive bowel management to keep them clean.

The surgeon should be able to predict in advance which children have good functional prognosis and which will not. This will help the surgeons in giving a realistic picture of the child's chances of bowel control. This will avoid creating false expectation and frustration later on among parents(table 18) .

Table 18: Common indicators of good and poor prognosis

Good prognosis signs	Bad prognosis signs
Good bowel movement pattern: 1-2 bowel movements per day, no soiling in between	Constant soiling and passing stool
Evidence of sensation when passing stool (pushing, making faeces)	No sensation, no pushing
Urinary control	Urinary incontinence, dribbling of urine

After the main repair and colostomy closure, it is possible to establish the functional prognosis. If the child's defect is of the type associated with good prognosis such as a vestibular fistula, perineal fistula, rectal fistula, rectourethral bulbar fistula, one should expect that the child would have voluntary bowel movements by the age of three years. These children will still need supervision to avoid fecal impaction, constipation and soiling. The child with poor prognosis, the BMP is needed to keep the child clean and it should be implemented when the child is 3 or 4 years. A child with a rectoprostatic fistula has ~50% chance of being continent. Therefore effort should be made to achieve toilet training by the age of 3 years.

If child with good prognosis type has been previously operated and has fecal incontinence, a redo posterior sagittal anorectoplasty (PSARP) can be performed and the rectum can be relocated within the limits of the sphincter mechanism. Approximately 50% of the children operated on under these very specific circumstances have a significant improvement in bowel control (117). [Table 19]

Table 19: Predictions of functional prognosis

Indicators of good prognosis for bowel control	Indicators of poor prognosis for bowel control
1. Normal sacrum 2. Prominent midline groove (good muscles) 3. Some types of anorectal malformations • Rectal atresia • Vestibular fistula • Imperforate anus without fistula • Cloacas with a common channel <3 cms • Less complex malfunctions: perineal fistula	1. Abnormal sacrum 2. Flat perineum (poor muscles) 3. Some types of anorectal malformations • Rectobladder neck fistula • Cloacas with a common channel >3 cms • Complex malformations

Children operated suffer from fecal incontinence can be divided into two well-defined groups that require individualized treatment plans: 1) those with constipation (colonic hypomotility). 2) children with loose stools and diarrhoea

1) Children with constipation (colonic hypomotility): In these the motility of colon is significantly reduced. Constipation is very helpful, as it guarantees that they will remain clean in between enemas. The basis of bowel management programme is to teach the parents to clean the child's colon once a day with suppository, an enema and colonic irrigations. The fact that they suffer from constipation is very helpful ,as it guarantees that they will remain clean in between enemas. No special diet or medications are necessary.

2) Children with loose stools and diarrhoea: The great majority of children who suffer from these are those in whom resection of rectosigmoid (reservoir) have been done (108, 118). Therefore, this group of children have overactive colons. Rapid transit of stool results in frequent

episodes of diarrhoea. This means that even when an enema cleans their colon rather easily, stool keeps passing fairly quickly from the cecum to the descending colon and the anus. To prevent this, a constipating diet and low medications to slow down the colon are necessary. Eliminating foods that further loosen bowel movements will help the colon to slow down.

A contrast enema study with hydrosoluble material will help in determining the type of colonic motility: hypomotility constipated or hypermotility. The bowel management program is then implemented according to the patients' type of colon and the results are evaluated every day.

Types of Enema :

There are different types of solutions that can be used for used for enemas: ready made enemas or solutions prepared at home based on salt and water (0.9% saline can be made by adding 3-4 teaspoons to 1 litre of water). Saline enemas are effective and less expensive. Phosphate enemas may cause abdominal cramps and can cause phosphate intoxication and children with impaired renal function should use with caution. Glycerine can be added to the saline enema to make it more effective. The frequency and the amount of enema also can be increased initially to get the desired effect. The "right" enema is the one that can empty the child's colon and allow him to stay clean for the following 24 hours. This can be achieved by trial and error and learning from previous attempts.

The child with the overactive bowel along with enema, needs constipation diet and drugs like loperamide are recommended. Most parents would eventually know which meals provoke diarrhoea and which constipates the child.

Every summer holidays, the children with some potential for bowel control can try to find out how well they can control their bowel movement without the help of enema. This is tried during vacations to avoid accidents at school.

Continent catheterisable stoma :

Most pre-school and school-age children enjoy a good quality of life while undergoing the bowel management program. However, when they reach puberty many express a high degree of dissatisfaction. They feel that their parents are intruding on their privacy by giving them enema. It is feasible for them to administer the enema by themselves. An operation called the continent appendicostomy or a Malone procedure has been designed for this group of specific children. Malone procedure is just another way to administer an enema and therefore before implementing the Malone procedure, the child had to be perfectly clean with his or her regular bowel management. The operation consists of connecting the appendix to the umbilicus and creating a valve mechanism that allows catheterisation of the appendix for the enema fluid, but avoids leakage of stool through it (119). The other advantages are easy access to the colon, one way effective irrigation and smaller volumes for irrigation and physiological comfort.

Continent cecostomy techniques :

1) Disconnection and reimplantation of the appendix .
2) Orthotopic appendicostomy (+/- divided appendix).
3) Tubularised cecal/colonic flap.
4) Transverse tubularised ileal tube (Monti).
5) Laproscopic MACE (appendicostomy only).
6) Cecostomy button.
7) Percutaneous cecostomy catheter.

8) LACE (left colonic antegrade colonic enema).

Dietary prevention of constipation :

Dietary fiber: Roughage of fibers plays an important role in the passage of chime. Roughage increases the digestive juices and it swells up in the intestine due to its absorption of water. It serves as culture medium for the bacteria in the colon. The breakdown of the fibers by bacteria creates gases and acids, which in turn stimulates the peristalsis of the intestinal wall. The consistency of the stool becomes softer and the distension of the intestinal wall and increased propulsive motility shortens the transit time and reduces the water reabsorption. For the roughage to have the optimal effect it is important to drink enough liquid. Roughage is indigestible vegetable material which can be found in leaves, fruits and roots. Non-purified vegetable fibers are the fibers found in the cereals, fruits and vegetables. Purified vegetable fibers are fibrous and polymer substances such as liquid cellulose and pectin. Food stuff containing a lot of fibers are fruits and vegetables especially berries, dried fruits, green peas, pulses, whole cereals, bran, nuts and almonds.

Dietary stimulants: In addition to the mechanical stimulation a chemical stimulation may also improve peristalsis . Lactic acid is an example and is found in yoghurt , butter milk ,pickles and vegetable juices .Tartaric acid found in grapes , apples, citric acid in citric fruits and acetic acid in wine , vinegar are believed to improve peristalsis .Lactase or concentrated sugar solution made of lactose also stimulate intestinal motility.

Operations to improve continence after previous surgery

Several techniques have been described to restore continence after previous repair of anorectal malformation. Secondary operations are done mainly in two groups of patients. The first being in patients with a benign type of defect like "recto- bulbar fistula" who is incontinent post-operatively due to totally misplaced rectum where the options are (1) redo PSARP (2) Stephens secondary pull-through. The second group is represented by patients who had previous operations but who suffer from fecal incontinence. In principle the attempts to correct this is based on：

(1) Secondary repair of levator ani – Stephen's secondary repair of damaged or hypoplastic muscle complex (120).

(2) Reinforcement and substitution for the levator ani – (a) Kottmeier's levatorplasty (121) (b) Puri & Nixon's levatorplasty (122).

(3) Free autogenous muscle transplant for strengthening of the levator ani (Palmaris Longus Transplant) (123).

(4) Substitution of striated anal sphincter (a) Gracilis muscle transplant (124); (b) Dynamic Graciloplasty; (c) Gluteus maximus transplant (125).

(5) Construction of sphincter from bowel wall (a) free smooth muscle transplantation; (b) flap smooth transplantation (126).

(6) Electric devices to stimulate the sphincters (127).

(7)Secondary procedures for anal prolapse or stricture (a) Nixon's anoplasty (128); (b) Mollard – Laberge operation (129)

Resection of inert rectostigmoid for the treatment of chronic constipation

Many children with chronic constipation after repair of ARM suffer from different degree of dilatation of rectum and sigmoid, a condition defined as mega-rectosigmoid, due to hypomotility disorder that interferes with complete emptying of the rectosigmoid. This is mainly due to inappropriate treatment for constipations, leading on to fecal impaction and overflow incontinence. The impaction is removed with enemas and/or colonic irrigation to clean the mega-rectosigmoid. The constipation is subsequently treated with large doses of laxatives. The dosage of laxative is increased till the child is able to completely empty the colon everyday. If the medical management is difficult and the dosage required to treat continent is very high, sigmoid resection may be beneficial. The very dilated mega-rectosigmoid is resected and the descending colon is anastomosed to the rectum. After resection, the amount of laxative can be minimized or eliminated. Pena et al followed up 53 cases after sigmoid resection. Following the resection 10% did not require laxative and the rest required significantly less amounts of laxative. It is found that the patients who benefit are these with localized forms of mega-rectosigmoid.

10. Electromanometric Evaluation

For the complete evaluation of postoperative continence, anorectal manometric studies have been performed on patients with ARM.(130,131).

Manometry Study: Manometry is done without anesthesia ,except in restless children ,who required mild sedation at the time of examination. The probe is filled with water before examination, but is not perfused during the test. The anorectal pressure profile is first recorded in centimetres by withdrawing the probe that is introduced 8cms above the mucocutaneous line into the rectum. The presence or absence of anorectal reflex is determined by distending the balloon in the rectum.

Anal manometry has been used to measure anorectal resting & squeeze pressure profile, the rectal adaptive reaction & the internal sphincter relaxation & the anorectal reflex. . According to Holschneider (132) the normal findings seen postoperatively after pull-through were (1) anorectal pressure profile 25+/-5mmHg. (2) squeeze pressure profile of more than 35mmHg (3). A normal rectal adaptation reaction (4) Normal rectal internal sphincter relaxation.

Patients with a good clinical result after staged abdominoperineal rectoplasty or perineoplasty exhibited the same anorectal pressure profile as normal subjects, with a high pressure zone in the anal canal (133) . They also have an adequate anorectal pressure difference that is not significantly different from that of normal subjects (133).On the other hand patients with a poor clinical result showed a slight radial change in the anorectal pressure profile and did not have such a high pressure zone as was found in normal subjects (33) .Most of the patients with a good clinical results regardless of the type of anomaly exhibited anorectal reflex . In high anomaly however some patients with a good clinical result did not show the reflex (133).

Manometeric investigations showed that good clinical results after surgery were associated with

normal function of the anorectum. The anorectal pressure profile observed in all patients with adequate continence characteristically had a marked high pressure zone as the normal subjects. Thus the presence of a normal anal pressure at rest as well as adequate anorectal pressure difference was found to correlate well with continence after surgery . However the anorectal reflex in high anomalies did not necessarily correlate well with continence . Accordingly it seems that the reflex is not essential to achieve continence in patients with high anomalies . This might be explained as follows : In high anomaly , only a mechanical resistance remains without sensitive receptors in the mucosa which is concerned with initiating anorectal reflex . Therefore normal anal resting pressure and an adequate anorectal pressure difference in a high anomaly are apparently more important factors related to continence after reconstructive surgery .

Improvement in bowel function with puberty :

Fecal incontinence in patients having undergone repair for ARM improves at adolescence , as constipation disappears (101) . This improvement with time is probably is related to reinforced sphincter function and an increasing use of gluteal and pelvic floor muscle and is a manifestation of the adaptation and adjustment made by the patient himself to achieve socially acceptable status .

11. Magnetic Resonance Imaging (MRI)

Endoanal ultrasound, CT scan and MRI provide excellent information about the post operative anatomy of the ARM. Hence imaging studies are used to assess long term anorectal function in patients with ARM .

Pelvic MRI is a useful tool for assessment of anorectal malformation before and after the initial repair (134). Advantages include excellent inherent soft tissue contrast enhancement, multiplanar imaging capability and lack of ionizing radiation .Disadvantages include cost, the relatively frequent need for sedation and a lack of access to the technique in some locations.

Pelvic MRI of patients who have undergone surgical repair of ARM is performed with high resolution phased – array coils such as eight channel cardiac or torso phased array coils .The imaging protocol includes T-1 and fast or turbo spin – echo T2 weighted sequence in the axial , sagittal and coronal planes .To highlight the low-signal-intensity fat and mucosa , fat saturation is not used .The surgeon is interested in the mid sagittal section because its the plane used for the operative approach .An optional sequence is oblique coronal T2 weighted images angulated in line with anal canal when for further clarification of the sphincter – bowel relation is necessary (135). Axial T2 weighted images with fat suppression may be helpful for differentiating associated anomalies of the lower genitourinary tract (135).

Children with anorectal malformation have variable degrees of striated muscle development from near normal muscle to complete absence of the sphincter muscle (136).The striated muscle component of the sphincter mechanism is well assessed with MRI .Assessment of muscle quality is subjective and based on internal comparison for symmetry , comparison with pelvic MRI of healthy persons and ease of difficulty of visualisation of the muscle in the different planes . The sphincter muscle complex is best seen on axial images at the level of the symphysis pubis and below (137). Coronal and sagittal images are necessary to verify findings on the axial images and to assist in

ascertaining the location of the bowel in relation to the muscle . After identification of the muscles of the sphincter mechanism, it is important to assess the relation between the sphincter and the pulled through bowel . Malpositioning of the rectal pull through can be identified on axial, coronal and sagittal planes. Axial and coronal images best show side to side displacement of the bowel .Sagittal images help in assessment of anterioposterior displacement of the bowel in relation to the sphincter .The most commonly reported error is anterior misplacement of the pull - through bowel within the sphincter (138). In some cases , the bowel is properly located , but mesenteric fat inadvertently pulled with bowel through the sphincter during the initial repair interferes with the continence (134). Axial T-1 and T-2 weighted images without fat saturation shows this fat as a halo of high signal intensity surrounding the wall of the pull through bowel. .

Fukuya et al (139) found that MRI evaluation based solely on muscle development can be misleading and therefore they included a measurement of the anorectal angle in their postoperative evaluation and formed a statistically significant difference in anorectal angle between patients with good and poor clinical outcome Anorectal angulation represents sufficient contraction of the sphincter muscle in the postoperative period .

de Souza et al (140) used a qualitative index of sphincter appearance .A sphincter that appeared normal was scored 0 , minimal asymmetry of the muscle deemed a mild deficiency and scored 1, a 25-50% reduction in the length /thickness of any sphincter component compared to the other side was a moderate deficiency and scored 2 and such reduction that was greater than 50% was deemed severely deficient and scored 3 . Tang et al (141) measured anorectal muscle complex in normal children using phase – arrayed MRI and found that absolute measurement values of the muscle cannot be compared among children of different ages .So he used a ratio between the absolute width of the muscle and the transverse and anteroposterior diameter of the pelvis as the relative width of the muscle to avoid the influence of age . The dimensions of the puborectalis muscle and external anal sphincter were measured in different planes . Normal relative length of the

puborectalis and external anal sphincter were measured as 0.47+/-0.04 and 0.41+/-0.04, respectively, and the relative width of puborectalis and posterior external anal sphincter were 0.50+/-0.04 and 0.44+/-0.04 in children younger than 14 years.

Yutaka Sato et al (153) demonstrated MR imaging of normal musculature in various planes.

Among transverse images, two are particularly important : the plane through the symphysis pubis and coccyx and through the ischial rami . The plane through the symphysis pubis and coccyx (PC-plane) includes the prostate or cervix and puborectalis muscle .It corresponds to the pubococcygeal plane of the invertogram. At this level, the rectum lies immediately posterior to the prostate or cervix and is surrounded by the triangular shaped muscle of the puborectalis muscle with its apex directed posteriorly . At the same level, the gluteus maximus muscle approximates each other in the midline over the coccyx. The plane through the ischial rami (I-plane) corresponds to the I-point plane of the invertogram and includes the bulb of the penis and the external sphincter. This plane approximates the junction between the rectum and the anal canal in healthy subjects. The external anal sphincter is oval shaped with the longer axis directed anterioposteriorly .At this and all caudal levels, the rectum or anal canal lies in the center of the external anal sphincter . Paired superficial transverse perineal muscle defines the anterior border of the external anal sphincter and extends laterally to the medial aspect of the ischial rami . In the coronal plane of the posterior pelvis, the levator ani supports the pelvic floor and the rectum rests upon the levator hammock. In the mid-pelvic plane, the rectum and anal canal penetrate the external anal sphincter. The inferior portion of the puborectalis muscle and superior portion of the external anal sphincter are contiguous and inseparable. In the anterior pelvic plane , the urogenital diaphragm ,prostate, and penile bulb are visualised . In mid-sagittal plane, the sphincteric muscle appears as a curved band-like structure posterior to the prostate or vagina. Margins of the sphincteric muscles are not distinct because of its tapering edges. The tip of coccyx should reach the level of the symphysis pubis. In the parasagittal plane ,the combined illiococcygeal and pubococcygeal portion of the levator ani is seen as a curved

linear structure on which the rectal ampulla rests .

Pringle KC et al (154) reported the use of MRI as a tool to plan surgical procedures in patients with imperforate anus , imaging the pelvis and lumbosacral spine in the sagittal, transverse, and the coronal planes. MRI clearly reveals the extent of the pelvic musculature in patients even with sacral agenesis. MRI is extremely useful in assessing patients under consideration for re-operation, clearly demonstrating the relationship between the pulled through colon and the striated muscle complex. A very useful addition is the ability to, on the same study, to detect previously unsuspected anomalies such as tethered cord, lipoma of the filium terminale and renal dysplasia.

Todd M Sachs et al (155) reported the use of MRI in planning operative strategy and predicting outcome by providing information about pelvic musculature.

12. MATERIALS & METHODS

METHODOLOGY

Fifteen children with intermediate type of anorectal malformation had undergone sacro-perineal pull-through between 2006-2009 in the Department of Paediatric Surgery, Christian Medical College Vellore. All of these had accurate localisation of sphincter muscle complex using muscle stimulator at the time of surgery before muscle relaxant was given. Perineum was stimulated before skin incision was made and the point of maximum contraction was marked as the site of proposed anus. After exposing the bowel and the puborectalis through sacroperineal route, the puborectalis was stimulated to localise it accurately and passage was made through the centre of the muscle complex into the proposed anus site. This passage was dilated to bring bowel through it after dividing the fistula. Out of these fifteen children, nine responded for follow-up and included in the study.

The follow up period ranged from 6 to 18 months with a mean follow up period 12 months.

All the 9 children underwent clinical evaluation to assess their somatic growth & functional status with respect to fecal continence.

X-ray of the abdomen was done to assess the fecal load.

Associated anomalies were also noted.

All the 9 children with intermediate anorectal malformation had **MRI evaluation of the pelvis** to determine the relationship of the pulled – through bowel with the sphincter muscle complex. MRI examination was done using either 1.5 T or 3 T magnets. All subjects were positioned supine with the pelvis centered on the coil & a body phase array was used . Sagittal , coronal, and transverse T 2 weighted images of the pelvic region were obtained in all subjects. Slice thickness was kept at 3 mm with interslice gap of 0.3 . Anorectal angle & thickness of the sphincter muscle was also studied.

Anorectal manometry was done in eight children.. All the eight children were sedated with triclorofos. Balloon tip pressure transducer probes were used. Pressure was recorded at 1cm , 2cm, 3cm from the anal verge & the maximum value was selected. Afterwards a balloon tipped catheter was passed into the rectum and inflated and the ano-rectal inhibitory reflex was noted.

EXCLUSION CRITERIA :

1. Children with low type of anorectal malformation.

2. Children with intermediate or high type of anorectal malformation who were operated elsewhere and then presented for re-do surgery.

3. Children with neurological abnormalities like myelomeningocele , sacral agenesis which by itself affect the continence mechanism.

4. Children with associated Hirschsprung's disease.

SACROPERINEAL PULL-THROUGH

POSITION OF THE PATIENT FOR SACROPERINEAL-PULLTHROUGH

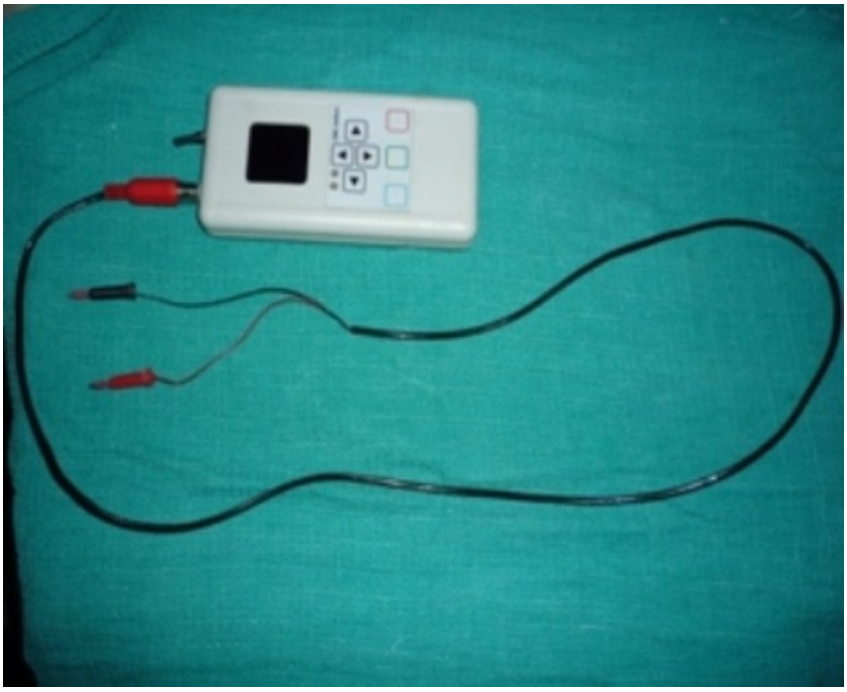
SPHINCTER MUSCLE COMPLEX IDENTIFIED WITH MUSCLE STIMULATOR

SACROPERINEAL INCISION BEING MADE

RECTUM BEING PULLED DOWN

-

ANOPLASTY COMPLETED



MUSCLE STIMULATOR

ANAL MANOMETRIC TRACING WITH ANAL STIMULATOR

13. RESULTS AND ANALYSIS

RESULTS AND ANALYSIS

A total of nine cases that had undergone sacroperineal pull-through were available for follow-up. The ages ranged from 5 months to 13 months with mean age range of 9 months .

DURATION OF FOLLOW UP

Follow up after colostomy closure ranged from 6 months to 18 months with mean follow up of 12 months.

ASSOCIATED ANOMALIES

Associated anomalies were seen in 5 of the 9 patients. 3 patients had grade 3 vesicoureteric reflux. 1 patient had undescended testis. 1 patient had mild hydroureteronephrosis. 2 patients had congenital heart disease, 1 with mitral regurgitation, mitral valve prolapse , and patent ductus arteriosus and the other had patent ductus arteriosus with atrial septal defect . 1 patient had Down's Syndrome.

HEIGHT AND WEIGHT

The height and weight were plotted against the growth curve graph recommended by the Growth monitoring Guidelines Consensus Meeting of Indian Academy of Paediatrics (142). The reference growth chart was applicable to the Indian population. 2/9 (22.2%) of the children were below 50th percentile for height and 7/9 (77.7 %) were below 50th percentile for weight (Table20).

Table 20: Growth pattern in children with anorectal malformation

Height (percentile)	No.	Weight (percentile)	No.
< 3	_	<3	4
3 - 25	_	3 -25	1
25 - 50	2	25 - 50	2
50 - 75	3	50 - 75	_
50 - 97	4	50 - 97	2

PERINEUM :

Position of the neoanus

The anus post operatively was in normal position in all patients.

Patency

4 of the 9 (44.4 %) had a supple and patent neoanus. The remaining 5 (55.5%) children had varying degrees of stricture. Out of these 5 children, 3 children were severe enough to require anoplasty. The remaining 2 children are managing with anal dilatation..

Mucosal Prolapse

All the 9 (100%) cases had mucosal prolapse . Of these 7 were major and underwent anal mucosal trimming.

Genitourinary anomalies

1. Vesico-ureteric reflux : 3 children had vesico-ureteric reflux. 2 had unilateral reflux and 1 bilateral reflux and all are being managed conservatively.

2. Undescended testis: 2 had unilateral undescended testis and both have undergone orchidopexy.

BOWEL FUNCTION

Mean age at follow up was 9 months and therefore it was too early to assess the bowel function using any objective scoring system as the children were not yet toilet trained.

However 7 mothers reported that their children were passing stools frequently in small amounts throughout the day. One child is passing stool about twice a day with the help of laxative. In one child colostomy closure was done very recently , hence bowel habit could not be assessed .

4) Fecal Load as seen on X – rays :

X-ray abdomen was taken in 8 patients to assess the fecal load. A score of 1 to 4 were given according to the extent of fecal matter present in the colon. Score of 1 was given if the fecal load was limited to sigmoid and descending colon, score of 2 if the fecal load extended up to the whole of transverse colon and score of 3 if the fecal load extended up to the caecum. Score of 4 was given if a megarectum or megasigmoid was evident .

Radiological extent of fecal load in the study is given in Table 21 below .

Table 21 : Radiological extent of fecal load

Fecal load extent	Grade	No. in the study
Sigmoid & descending colon	1	3 (37.5%)
Whole of Transverse colon	2	_
Up to Caecum	3	4 (50%)
Megasigmoid/ megarectum	4	_

According to the above scores, score of 1 was seen in 3 and score of 3 in 4 children..

Anal Manometry :

Anal manometry was done in 8 patients.

The average anal resting pressure was 63.3 cm of H_2O .

The rectoanal inhibitory reflex was present in 4 patients in our study.

Table 22: Anorectal pressure and rectoanal inhibitory reflex.

SL.NO.	HOSP. NO.	ANAL RESTING PRESS. (CM OF H_2O)	RECTOANAL INHIBITORY REFLEX
1.	006722D	66	+
2.	032014D	18	_
3	048013D	144	+
4	143263D	18	+
5.	056734D	108	+
6.	023601D	45	_
7	106889D	72	_
8.	904323C	36	_

Magnetic Resonance Imaging:

Magnetic resonance imaging was done in all the 9 children.

The position of the rectum at PC line, where bowel enters the levator ani complex was assessed.

Thickness of sphincter muscle complex at I-point, M-point and at the anal verge was measured.

The position of the rectum where it enters the puborectalis was central in all the 9 children. The measurements of the external sphincter thickness are given in Table 23. From this table it is obvious that the thickness of external sphincter is uniformly thin on the right side in all the patients. At the anal verge the sphincter thickness seems to be satisfactory in 5 patients, very thin on the right side in 1 patient and absent on the right side in 3 patients. These are diagrammatically shown in pages 80-82. The anorectal angle was clearly seen in the sagittal plane in 8 children. The average anorectal angle was 158.6 . In 3 children the angle was above 158 and in 5 children the angle was below 158. _

Table 23 : MRI Findings

SL. NO.	HOSP NO.						IMENT ON SPHINCTER
1	00672 D						emely thin on
2	03201 D						emely thin on
3	04801 D						emely thin on fat replaced
4	31046 D						emely thin on whole length
5	14326 D		right				cient on right 6'O – 12'O clock
6	056734 D	Central	Grossly thin bilaterally	R 0 L 0	R 0 L0	R 1.9 L 1.9	Atrophic thin strands
7	023601 D	Central	Mild focal thinning on right	R 0 L1.7	R 0 L 1.1	R 0 L 2.8	Thin irregular. ight wall fat replaced
8	106889 D	Central	Grossly thin on right	R 0 L 2.6	R 0 L 1.4	R 2 L 2.8	Right wall grossly thin
9	904323 C	Central	Bilateral thin right < left	R 0 L 3	R 0 L 3.2	R 4.2 L 3.6	Bilateral grossly thin

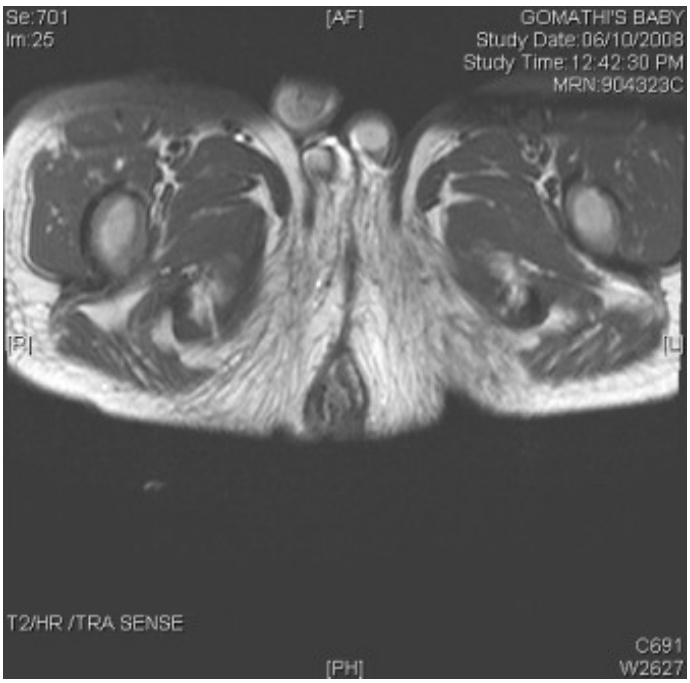
MAJOR MUCOSAL PROLAPSE WITH ANAL STENOSIS

MRI FINDINGS

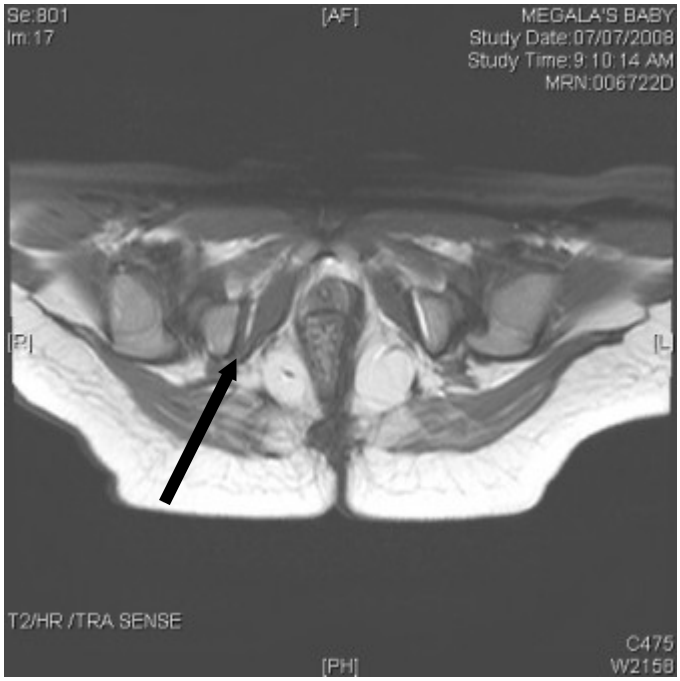
TRANSVERSE SECTION AT PC-LINE SHOWING RECTUM IN CENTRAL POSITION (thick black arrow)

MID-SAGITTAL SECTION SHOWING ANORECTAL ANGLE OF 155 DEGREES (thick white line)

MID-SAGITTAL SECTION SHOWING ANORECTAL ANGLE OF 180 DEGREES (thick white line)



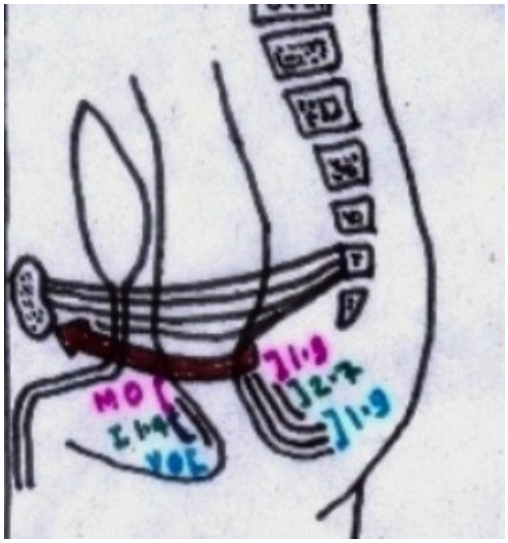
TRANSVERSE SECTION SHOWING EXTERNAL SPHINCTER AT THE LEVEL OF ANAL VERGE (thick black arrow)



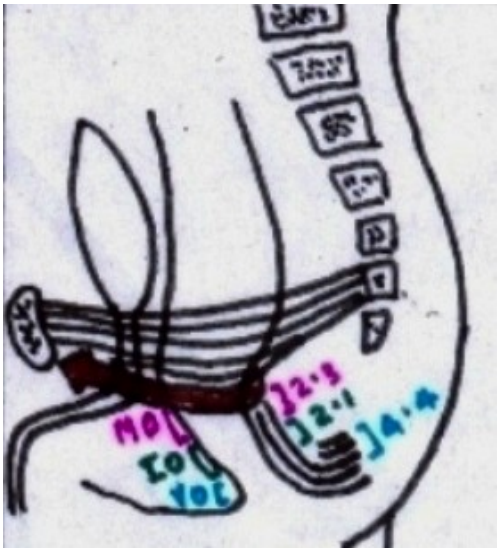
TRANSVERSE SECTION SHOWING EXTERNAL SPHINCTER AT THE LEVEL OF I – POINT (thick black arrow)

TRANSVERSE SECTION SHOWING EXTERNAL SPHINCTER AT THE LEVEL OF M – POINT (thick black arrow)

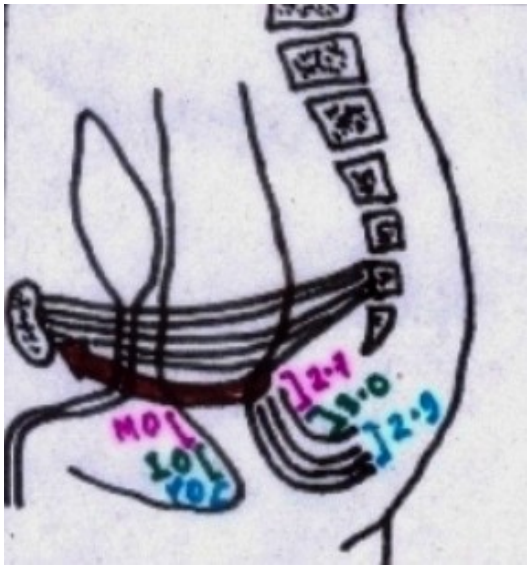
DIAGRAMATIC REPRESENTATION OF THICKNESS OF EXTERNAL SPHINCTER OF ALL NINE CASES.

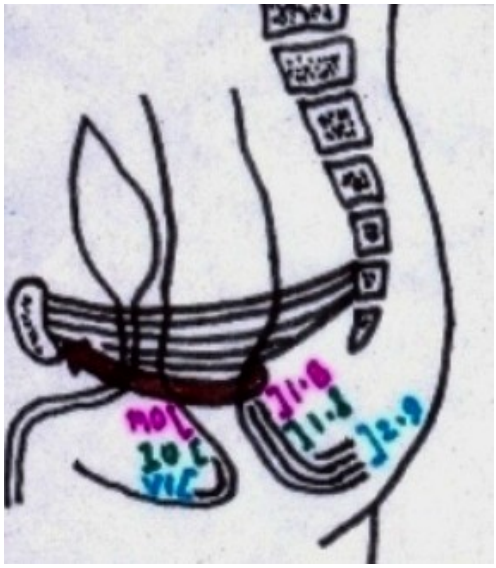


CASE – 1



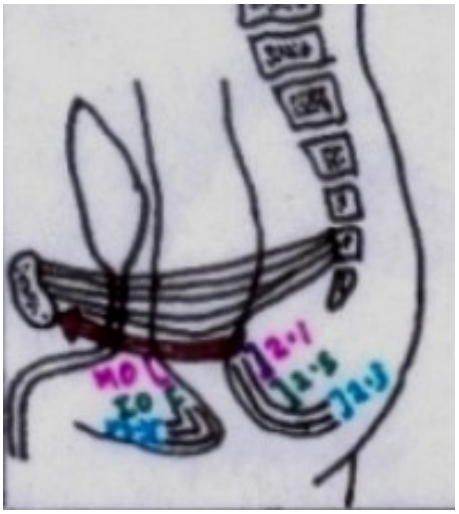
CASE – 2





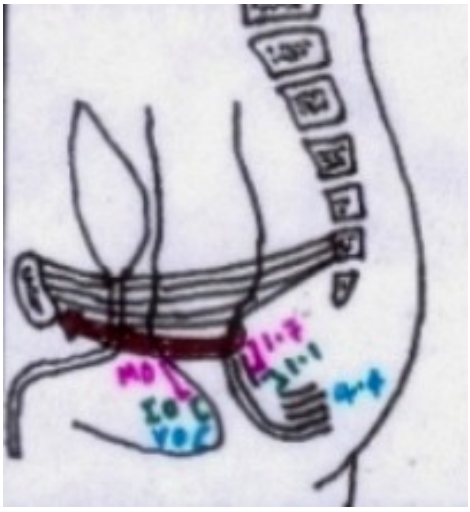
CASE – 3

CASE – 4

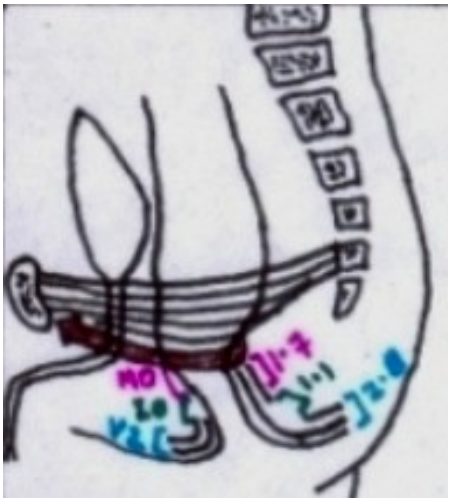


CASE – 5

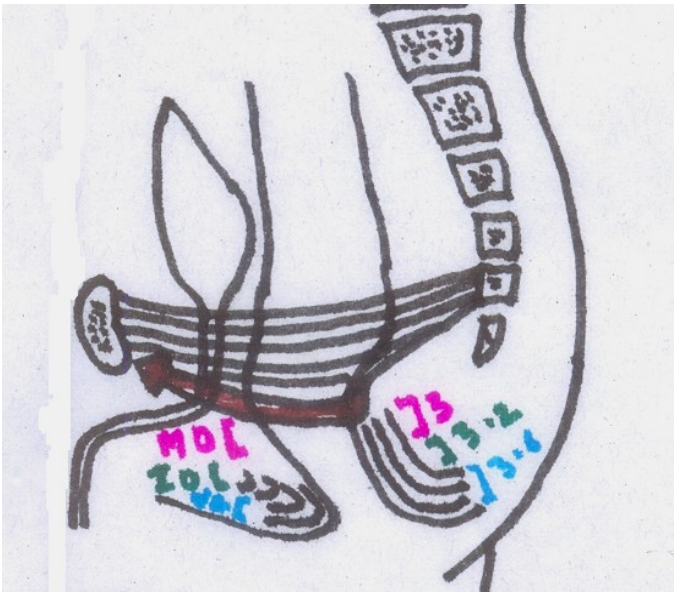
CASE – 6



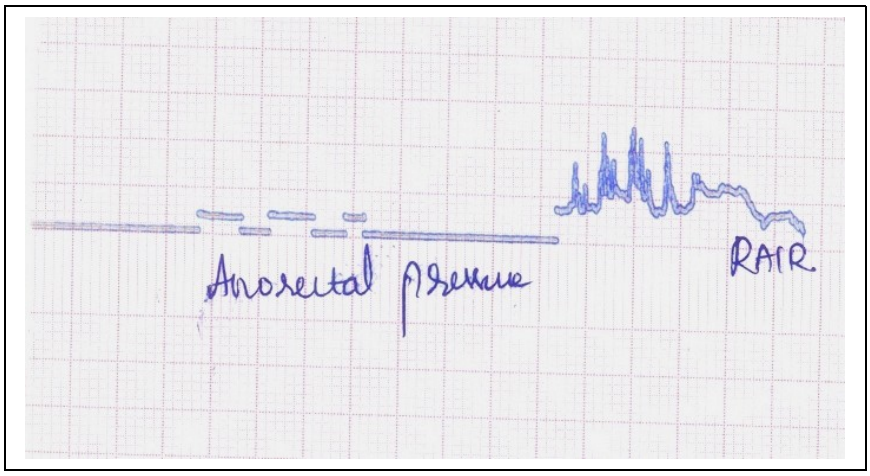
CASE – 7



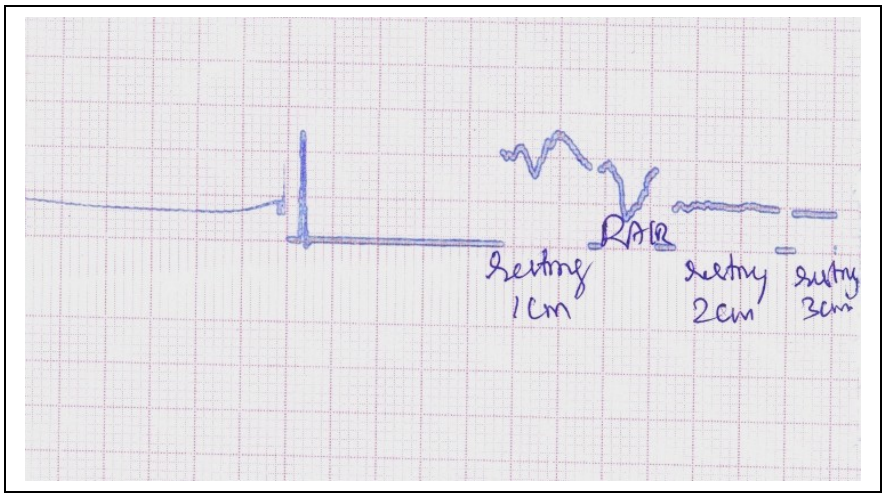
CASE – 8



CASE – 9



GRAPH SHOWING ANORECTAL PRESSURE PROFILE AT : LEVEL 1 : 9CM OF H_2O, LEVEL 2 : 18CM OF H_2O, LEVEL 3 :18 CM OF H_2O AND THE RECTOANAL INHIBITORY REFLEX.



GRAPH SHOWING ANORECTAL PRESSURE PROFILE AT : LEVEL 1: 108 CM OF H_2O, LEVEL 2: 46 CM OF H_2O ,LEVEL 3: 38 CM OF H_2O AND THE RECTOANAL INHIBITORY REFLEX.

14. DISCUSSION

DISCUSSION

This study was done in children who had undergone sacroperineal pull-through operations for intermediate anorectal malformation after intraoperative localisation of sphincter muscle complex with muscle stimulator in Christian Medical College , Vellore from 2006 to 2009. These children were followed up.

The somatic growth assessment of these children showed that.2/9 (22.2%) of children were below 50th percentile for height and 7/9 (77.7%) were below 50th percentile for weight. Studies on somatic growth pattern in patients on follow up of pull-through operation are lacking. In 13 patients (59.2%) the height & weight were less than the 50th percentile of the expected values for their age in a follow up study of pouch colon.(144). In our previous study " Follow up of children with ARM " done in our department showed that 55% were below 25th percentile for height and 75% were below 25th percentile for weight.

Perineum

The position of the neoanus was in normal position in all the patients. The anal opening had varying degrees of anal stenosis in 5/9 (55.5%). Out of these 5 patients 3 have undergone anoplasty for their stenosis . Major mucosal prolapse was in 7/9 patients (77.7%) and all these 7 patients underwent anal mucosal trimming for their prolapse. In literature the incidence of anal stenosis and anal mucosal prolapse ranges from 15-78% (145). In our study all our children required secondary anal procedures (anoplasty -3 and anal mucosal trimming -7) .Our tendency had been to leave the bowel protruding out of the anal verge, out of fear of retraction which would explain the high incidence of mucosal prolapse. Pena emphasizes suturing the bowel to the anal verge under mild tension and

accurate skin to mucosa approximation. Anal stenosis in our study was high because there was no strict anal dilatation program as suggested by Pena. Pena considers lack of anal dilatation as an important factor for anal stenosis and advices a strict dilatation program starting 2 weeks after repair. The parents are advised to dilate daily twice and continue dilatation beyond closure colostomy according to the following protocol : once a day for 1 month, every 3rd day for a month, twice a week for a month, once a week for 1 month & once a month for 3 months (146). Constipation and soiling are major problems following repair of anorectal malformation.

Constipation was seen in 59% of bulbar fistula, 55% in imperforate anus without fistula and 45% of prostatic fistula on follow up in a series of 1192 patients by Pena (116) which was comparable to his early results (71) of 387 patients followed up which showed constipation in 55.5% of bulbar fistula, 50% of imperforate anus with no fistula, 41.4% of prostatic fistula and 25% of vaginal fistula. Another series by Chau-Jing (147) had constipation in 21% of bulbar and 17% of prostatic fistula. Constipation is the most common sequelae after ARM (116). Children in our study were too young to assess their bowel function. However there was even at this early age, tendency for constipation and fecal loading as seen in X-ray abdomen (87.7%). It is highly likely that these children will become constipated as they grow old .

Although it is too early to assess soiling in our children, seven mothers reported that their children are passing small amounts of stool very frequently throughout the day, which could develop into major soiling later on.

In our previous study “Follow-up of children with ARM” done in our department we have shown that soiling is the result of constipation and this can be managed by giving glycerine enema once daily in the morning .

Anal Manometry :

Holschneider reported that the normal findings seen postoperatively after pull-through were 1) anorectal pressure profile of 25 +/- 5 mmHg . 2) squeeze pressure profile of more than 35 mmHg . 3) a normal rectal adaptation reaction. 4) normal internal sphincter relaxation. (132).

It is established that the anorectal pressure profile observed in all patients with adequate continence characteristically has a marked high pressure zone. So presence of normal anal pressure profile at rest is essential for development of future continence (152). This high pressure zone probably reflects sphincter activity.

In our study 6/8 (75%) children had resting pressure above 47 mmHg, so it is possible that these children have good sphincter activity and will be continent in future.

Although squeeze pressure is an indicator of sphincter activity, it could not be assessed in our study as the children were too young.

The presence of internal sphincter in the pulled through bowel of ARM is disputed. Rintala observed good continence in children with positive internal anal sphincter relaxation and a high anorectal pressure profile in patients with internal sphincter preserving repairs (149). Presence of anorectal inhibitory reflex indicates the function of internal sphincter. Sangkhathat by studying 24 children who had ARM came to the conclusion that rectoanal inhibitory reflex plays a crucial role in emptying function after anoplasty and that these functions should be preserved during reconstruction.

In our study 4/8 (50%) children had showed positive anorectal inhibitory reflex even at this early age and so it is hoped that they will develop normal emptying function of the rectum in the future . We have tried to preserve as much of the rectal fistula as possible and to bring this end down.

MRI Findings :

In a study done by Hisato Tsuji et al (151) , the rectum was mispositioned in 21% in PSARP group and 25% of controls who had conventional surgery. In our study the bowel was found entering the puborectalis sling exactly in the centre in all patients 9/9 (100%).

Further down at the M-point and at I-point , it appears that the bowel is placed more eccentrically to the right.

Still further down at the anal verge , there is hardly any external sphincter on the right side in 4/9 (44.4%) children . Therefore it appears that although the bowel enters the centre of puborectalis sling , further down in the sphincter muscle complex, the bowel tends to be more towards the right . This could be either because of technical problem during placement of the bowel or due to sphincter-muscle hypoplasia.

It remains to be seen whether this finding will affect the ultimate bowel continence.

It has been shown by anorectal manometry that 6/8 (75%) children assessed have anal resting pressure above 47 mmHg. Only in 2/8 (25%) it was 13.2 mmHg. This indicates that there is evidence of sphincter muscle activity and it is to be seen in further follow-up, whether slight eccentricity of the bowel will actually affect the continence.

In 2 children where the anal resting pressure was below the normal , the external sphincter shown to be quite deficient on the right side as found in the MRI imaging..

The significance of these findings in relation to ultimate continence can be assessed only by following these children beyond puberty..

In our previous study “A Study of Children with Intermediate Anorectal Malformation who have undergone sacroperineal pull-through with regard to bowel function, quality of life and parental stress” where cases operated by sacroperineal technique without muscle stimulation the pulled through bowel was found to be eccentrically placed in 3/13 (43.3%) . But with muscle stimulation

in our present study the bowel has been within the sphincter muscle complex in all the 9 children.

In a study done by de Souza et al (140) it was found that in all children who had undergone PSARP reconstruction, there is a posterior midline linear scar which disrupts the sphincteric ring. Muscle which heals by fibrosis loses its elastic properties which may affect the continence mechanism. By our technique there is no disruption of the sphincter complex, because the sphincter itself is not divided. Hence this is a definite advantage over PSARP technique .

15. CONCLUSION

CONCLUSION :

- From our study it is found that with intraoperative localisation of the sphincter muscle complex with muscle stimulator, the bowel can be placed in the centre of the puborectalis sling without dividing the sphincter complex, thus avoiding fibrosis and damage to the sphincter muscle complex.

- As a further refinement to our technique, we hope to bring the bowel exactly in the centre of the external sphincter by passing initially a fine needle through the centre of the external sphincter identified precisely by the muscle stimulator, as a guide and then dilating it with a dilator and bringing the bowel down in the centre of the external sphincter.

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17. APPENDIX

PROFORMA

A. Patient details at first admission :

1. Name :
2. Age :
3. Hospital Number :
4. Address & Contact number:

5. Type of anomaly :
    a. Invertogram :

    b. Distal Cologram :

    c . Operative findings :

6 . Colostomy :
    a. Level :

7. Procedure:
    a . Age:
    b .Type of procedure:

8 .Complications:

a. Retraction:

b. Stenosis:

c.Prolapse:

d.Urinary:

9.Secondary Procedure:

10.Age of colostomy closure :

11.Associated Anomaly :

a.Cardiac:

b.Vertebral:

c.GIT:

d.Limbs:

e.Renal:

f.Miscellaneous:

B. Details of follow up :

1. Duration of follow-up from last surgery (colostomy closure)

2.Height (in percentile):

3.Weight(in percentile):

D.1.X-ray (to find fecal load):

2. Per-rectal examination for loaded colon:

3. Toilet trained or not:

4. Episode of enterocolitis (diarrhoea, fever, abdominal distention, falling sick)

E. Physical findings:

1.Examination of perineum:

a.Position:

b.Patency:

c.Prolapse:

d.Fistula:

e.Shelving:

2. Muscle stimulation:

3. Anal resting pressure profile and rectoanal inhibitory reflex :

4. MRI:

F. Urinary problems:

a. Dribbling:

b. Retention:

c. Recurrent UTI:

d. Structural abnormality:

G. Follow-up bowel management:

a.Enema type/frequency:

b.Duration of follow-up:

c.Results – functional score comparison:

MASTER CHART

SL. NO	NAME	HOSP. NO.	AGE	SEX	ASS. ANOMALY	HEIGHT	PERCENT	WEIGHT	PERCENT
1	Megala's baby	006722D	9 mo	M	LT. UDT PDA, ASD	90cms	97th	7.8kgs	3rd
2	Jothi's baby	032014D	6 mo	M	B/L GR 3 VUR	72cms	75th	7kgs	3rd
3	Padmasree's baby	048013D	9 mo	M	MR, MVP, PDA	73.5cms	90th	6.3kg	<3rd
4	Avirup Nandi	310461D	8 mo	M	-	68cms	50th	8kgs	50th
5	Sivakalai's baby	143263D	9 mo	M	LT. UDT LT. GR 3 VUR	71.5cms	75th	7.5kgs	10th
6	Urnav Gupta	056734D	6.5 mo	M	-	82cms	97th	12kgs	97th
7	Mala's baby	023601D	8 mo	M	LT.GR 3 VUR, LT. HUN	75.5cms	50th	7.25kgs	3rd
8	Sandip Soren	106889D	13mo	M	-	76cms	97th	9.3kgs	75th
9	Gomathy's baby	904323C	5 mo	M	-	100cms	97th	9kgs	50th

FOLLOW UP	SOILING	CONST-PATION	FECAL LOAD	ANAL POSITION	ANAL PATENCY	MUC. PROL	ANAL PROC	ANAL R PRESS.	ARIR
18 mo	3	-	Upto cecum	N	Patent	Major	Mucosal trimming	66	+
10 mo	2	-	-	N	Patent	Major	Mucosal trimming	18	_
18 mo	2	3	Upto cecum	N	Mild stenosis	Mild	-	144	+
8 mo	-	-	-	N	Mild stenosis	Mild	Anoplasty ,mucosal trimming	_	_
6 mo	2	-	-	N	Patent	Mild	Mucosal trimming	18	+
12 mo	3	3	Upto cecum	N	Major	Major	Mucosal trimming	108	+
11 mo	-	2	In sig. & rectum	N	Mild	Mild	Anoplasty mucosal trimming	45	_
9 mo	3	3	Upto cecum	N	Patent	Mild	-	72	_
12 mo	2	1	Upto sigmoid colon	N	Patent	Mild	Mucosal trimming	36	_

SL. NO.	HOSP. NO.	POSITION OF RECTUM (PC LINE)	COMMENT ON PRS	EXT. SPHINCTER MEASUREMENTS M pt. I pt. Anal verge	COMMENT ON EXT. SPHINCTER
1	006722D	Central	Asymmetric ; thin on left.	R 0 R 1.4 R 0 L 1.9 L 2.7 L 1.9	Extremely thin on right
2	032014D	Central	Extremely thin ;thinner on left	R 0 R 0 R 0 L 2.3 L2.1 L 4.4	Extremely thin on right
3	048013D	Central	Thin	R 0 R 0 R 0.9 L 2.7 L 3 L 2.9	Extremely thin on right ;fat replaced
4	310461D	Central	Asymmetric ; extremely thin on right	R0 R 0 R 1 L 1.8 L 1.8 L 2.9	Extremely thin on right whole length
5	143263D	Central	Totally deficient on right	R 0 R 0 R 3.3 L 2.1 L2.5 L 2.3	Deficient on right from 6'O – 12'O clock
6	056734D	Central	Grossly thin bilaterally	R 0 R 0 R 1.9 L 0 L0 L 1.9	Atrophic thin strands
7	023601D	Central	Mild focal thinning on right	R 0 R 0 R 0 L1.7 L 1.1 L 2.8	Thin irregular . right wall fat replaced
8	106889D	Central	Grossly thin on right	R 0 R 0 R 2 L 2.6 L 1.4 L 2.8	Right wall grossly thin
9	904323C	Central	Bilateral thin right < left	R 0 R 0 R 4.2 L 3 L 3.2 L 3.6	Bilateral grossly thin